# **Complete Summary**

#### **GUIDELINE TITLE**

Recommendations for physical activity and recreational sports participation for young patients with genetic cardiovascular disease.

# **BIBLIOGRAPHIC SOURCE(S)**

Maron BJ, Chaitman BR, Ackerman MJ, Bayes de Luna A, Corrado D, Crosson JE, Deal BJ, Driscoll DJ, Estes NA 3rd, Araujo CG, Liang DH, Mitten MJ, Myerburg RJ, Pelliccia A, Thompson PD, Towbin JA, Van Camp SP. Recommendations for physical activity and recreational sports participation for young patients with genetic cardiovascular diseases. Circulation 2004 Jun 8;109(22):2807-16. [86 references] PubMed

## **GUIDELINE STATUS**

This is the current release of the guideline.

## **COMPLETE SUMMARY CONTENT**

SCOPE

METHODOLOGY - including Rating Scheme and Cost Analysis RECOMMENDATIONS

EVIDENCE SUPPORTING THE RECOMMENDATIONS

BENEFITS/HARMS OF IMPLEMENTING THE GUIDELINE RECOMMENDATIONS OUALIFYING STATEMENTS

IMPLEMENTATION OF THE GUIDELINE

INSTITUTE OF MEDICINE (IOM) NATIONAL HEALTHCARE QUALITY REPORT CATEGORIES

IDENTIFYING INFORMATION AND AVAILABILITY

DISCLAIMER

#### **SCOPE**

# **DISEASE/CONDITION(S)**

Genetic cardiovascular diseases (GCVDs) including hypertrophic cardiomyopathy (HCM), arrhythmogenic right ventricular cardiomyopathy (ARVC), Marfan syndrome, and the ion channel diseases, including long-QT syndrome (LQTS), Brugada syndrome, and catecholaminergic polymorphic ventricular tachycardia (CPVT)

**Note**: Other uncommon familial conditions such as dilated cardiomyopathy or certain congenital heart malformations such as atrial or ventricular septal defect and mitral valve prolapse have not been specifically addressed here. However, it is possible to draw reasonable inferences from the recommendations presented and to extrapolate generally to these and other conditions.

## **GUIDELINE CATEGORY**

Management

#### **CLINICAL SPECIALTY**

Cardiology
Family Practice
Internal Medicine
Nursing
Pediatrics
Physical Medicine and Rehabilitation
Preventive Medicine
Sports Medicine

#### **INTENDED USERS**

Health Care Providers Physicians

# **GUIDELINE OBJECTIVE(S)**

- To develop consensus recommendations governing recreational exercise for patients with known genetic cardiovascular diseases (GCVDs)
- To facilitate the provision of high-quality patient care, to provide guidance to physicians, and to enable clinicians to base recommendations regarding medically acceptable recreational exercise programs for patients with known GCVDs on more than their own personal expertise and experience

## **TARGET POPULATION**

Young people (<40 years of age) with those inherited and nonischemic cardiovascular diseases implicated most often in sudden cardiac death in young people, and specifically for hypertrophic cardiomyopathy (HCM), the ion-channel disease (long QT-syndrome [LQTS] and Brugada syndrome), arrhythmogenic right ventricular cardiomyopathy (ARVC), and diseases of connective tissue such as Marfan syndrome (and related vascular conditions, Ehlers-Danlos syndrome, and other fibrillin disorders)

**Note**: These recommendations are not intended for individuals with the following clinical features: history of important cardiac symptoms including syncope or other important episodes of impaired consciousness; prior cardiac operation (including surgical septal myectomy for obstructive hypertrophic cardiomyopathy [HCM] and aortic root reconstruction for Marfan syndrome) or heart transplantation; presence of an implanted cardioverter-defibrillator or pacemaker; and clinically overt and potentially life-threatening arrhythmias or other evidence of high-risk status. The presence of any of these features may require individual clinical judgment in adapting the present exercise recommendations.

## INTERVENTIONS AND PRACTICES CONSIDERED

Patient education regarding appropriate recreational physical activity and exercise

**Note**: Recommendations for physical activity associated with performing art forms such as dance and ballet were not included.

## **MAJOR OUTCOMES CONSIDERED**

- Sudden cardiac death
- Maximum heart rate
- Metabolic equivalents

## **METHODOLOGY**

# METHODS USED TO COLLECT/SELECT EVIDENCE

Searches of Electronic Databases

## DESCRIPTION OF METHODS USED TO COLLECT/SELECT THE EVIDENCE

Not stated

#### NUMBER OF SOURCE DOCUMENTS

Not stated

# METHODS USED TO ASSESS THE QUALITY AND STRENGTH OF THE EVIDENCE

**Expert Consensus** 

## RATING SCHEME FOR THE STRENGTH OF THE EVIDENCE

Not applicable

#### METHODS USED TO ANALYZE THE EVIDENCE

Review

## **DESCRIPTION OF THE METHODS USED TO ANALYZE THE EVIDENCE**

Not stated

## METHODS USED TO FORMULATE THE RECOMMENDATIONS

**Expert Consensus** 

# DESCRIPTION OF METHODS USED TO FORMULATE THE RECOMMENDATIONS

The present statement represents the consensus of an international panel appointed by the American Heart Association Scientific Advisory and Coordinating Committee comprising clinical cardiovascular specialists and molecular biologists. This group has extensive experience with athletes of all ages but special expertise on the relationship between exercise and cardiovascular disease. The panel

deliberated on the benefits and risks of recreational exercise in patients with genetic cardiovascular disease (GCVD) and in this document has formulated prudent, contemporary, and practical consensus recommendations for physical activity in this group of cardiac patients.

In formulating the present consensus recommendations, the expert panel considered their individual and collective experiences, as well as available and pertinent scientific data. However, panel members were required to confront areas in which there is a paucity of precise published evidence. Indeed, the panel encountered few absolutes or truly quantitative data that could be used to formulate recommendations regarding exercise programs, physical activity, or lifestyle in noncompetitive recreational situations.

Given these limitations, the panel has nevertheless endeavored to offer recommendations that can be regarded as a reasonable, practical, and conservative framework for practicing physicians in advising patients with genetic cardiovascular diseases (GCVDs).

Finally, it was not the objective of the panel to restrict all exercise that could conceivably be associated with some increased risk but to develop a prudent balance between risk and benefit. Given the relative paucity of evidence in this area of medicine, particularly stringent recommendations would potentially (and unnecessarily) exclude a large proportion of patients with GCVD and in the process create a sedentary cardiac population deprived of the many benefits afforded by exercise for cardiovascular health.

## RATING SCHEME FOR THE STRENGTH OF THE RECOMMENDATIONS

Not applicable

## **COST ANALYSIS**

A formal cost analysis was not performed and published cost analyses were not reviewed.

#### METHOD OF GUIDELINE VALIDATION

Peer Review

#### **DESCRIPTION OF METHOD OF GUIDELINE VALIDATION**

This statement was approved by the American Heart Association Science Advisory and Coordinating Committee on March 26, 2004.

## **RECOMMENDATIONS**

## **MAJOR RECOMMENDATIONS**

## **General Principles**

The conveyance of exercise and lifestyle recommendations to young patients with genetic cardiovascular diseases (GCVD) requires substantial physician-patient interaction to ensure that recommendations are translated in sufficient detail. On the other hand, arbitrary and rigid directives would be impractical and most likely ineffective.

In communicating with patients, it is useful to define informal recreational sports by contrasting such activities with competitive sports. Because of the unique structure and pressures of organized sports, athletes with heart disease engaged in competition may not always use proper judgment in prudently extricating themselves immediately from vigorous exercise, even should they recognize the potential medical need to terminate the activity. For example, dizziness, palpitations, fatigue, excessive dyspnea, or chest discomfort (or any other potential warning sign of cardiovascular disease) experienced during competitive sports may be difficult for the participant to distinguish reliably from those innocent sensations that can normally accompany intense and extreme exercise and mimic symptoms of cardiac disease.

However, such considerations are not generally part of truly recreational sports activity, constituting a clear distinction on which exercise recommendations can be effectively conveyed to patients with GCVD. Participants in recreational physical activities have greater opportunity to exert reasonable control over their level of exercise and therefore are more likely to reliably detect cardiac symptoms and willfully terminate physical activity. On the other hand, some recreational sports (e.g., soccer, tennis, squash, and racquetball) can become truly competitive, largely owing to the style of play and attitudes of the participants, and thereby may obscure these important distinctions.

The panel has not included within the definition of recreational sports those neighborhood and elementary school activities for young children that involve lesser degrees of physical intensity and which should be an allowed component of daily activity for individuals with GCVD. Of note, some mildly symptomatic GCVD patients may be under the misconception that their functional limitation can be overcome by physical activity, rather than regarding symptoms provoked by exercise as "warning signs" triggered by their underlying heart disease.

#### **Patient Recommendations**

Sports activities have been categorized with regard to the high, moderate, and low levels of physical intensity required (see table below titled "Recommendations for the Acceptability of Recreational [Noncompetitive] Sports Activities and Exercise in patients with GCVDs"). These 3 partitions relate to the generally expected degree of physiological exertion implied by a sporting discipline. Patients with GCVD can safely participate in most forms of recreational exercise judged to be of moderate or low intensity (equivalent to 4 to 6 and <4 metabolic equivalents, respectively; Table).

Eligibility for exercise in specific recreational sports activities was assessed on a graded scale (from 0 to 5), with 0 to 1 designating activities generally not advised or strongly discouraged, 4 to 5 indicating activities probably permitted, and 2 to 3 indicating intermediate activities, which should be assessed on an individual basis. The assigned grades represent only an estimate, which assumes the usual level of

physical exertion for a given recreational activity. However, in large measure, this grading system cannot take into account other potentially important variables such as the psychological burden and physical intensity uniquely brought to a sport by an individual participant or the potential effects of cardioactive drugs, environmental conditions, and the precise clinical profile. Therefore, these recommendations are, to a certain extent, necessarily subjective and represent only a starting point for clinical judgments in individual asymptomatic (or only very mildly symptomatic) patients with clinically evident GCVD. Consequently, their application requires substantial reliance on the practice of the "art of medicine" and the weighing of perceived risk with respect to benefit for each patient.

Furthermore, these recommendations are not intended for individuals with the following clinical features: history of important cardiac symptoms including syncope or other important episodes of impaired consciousness; prior cardiac operation (including surgical septal myectomy for obstructive hypertrophic cardiomyopathy [HCM] and aortic root reconstruction for Marfan syndrome) or heart transplantation; presence of an implanted cardioverter-defibrillator or pacemaker; and clinically overt and potentially life-threatening arrhythmias or other evidence of high-risk status. The presence of any of these features may require individual clinical judgment in adapting the present exercise recommendations.

#### Activities That Should Be Avoided

The panel also found it useful to express specific exercise recommendations in terms of those activities that should be avoided by patients with clinically diagnosed GCVD:

- "Burst" exertion (or sprinting), characterized by rapid acceleration and deceleration over short distances. Exercise of this type is encountered in a variety of sports, such as basketball (particularly full-court play), soccer, and tennis. Therefore, preference is given to recreational sporting activities such as informal jogging without a training regimen, biking on level terrain, or lap swimming, in which energy expenditure is largely stable and consistent, even over relatively long distances or periods of time.
- Extremely adverse environmental conditions, which may be associated with alterations in blood volume, electrolytes, and state of hydration and thereby increase risk, such as greatly elevated or particularly cold temperatures disproportionate to that which the athlete is accustomed to in temperate climates (i.e., >80 degrees F [27 degrees C] and <32 degrees F [0 degrees C]), high humidity, or substantial altitude.
- Exercise programs (even if recreational in nature) that require systematic and progressive levels of exertion and are focused on achieving higher levels of conditioning and excellence, as in road running, cycling, and rowing. Patients with GCVDs such as HCM, in which limiting dyspnea may occur with exercise, should be discouraged from any exertion that provokes these symptoms. These individuals are also advised against systematic training during which they are extended beyond the physical limits imposed by their underlying disease and the average aerobic state expected at that age.
- Excessive participation in sporting activities that otherwise would be regarded as recreational if performed in moderation (e.g., downhill skiing continuously

- over an entire day versus more limited and selective skiing over the same time period).
- Exercise-related and adrenergic-type activities or stress that conveys a risk for cardiac events, specific to certain disease states. For example, in long-QT syndrome (LOTS), swimming, abrupt loud noises (such as from a race starter's pistol), and diving have been implicated as triggers for sudden death, particularly with certain mutant genes (i.e., KCNQ1 [or LQT1] for swimming and KCNH2/HERG [or LOT2] for auditory triggers). However, such laboratory-based molecular information is unlikely to be available to clinicians prospectively making exercise recommendations. Patients with rare conditions such as catecholaminergic polymorphic ventricular tachycardia (CPVT), in which many forms of exercise are associated with catecholamine release that triggers ventricular tachycardia, should be cautioned against virtually all forms of vigorous physical activity. The same restriction should be adopted for that subgroup of arrhythmogenic right ventricular cardiomyopathy (ARVC) patients that shares with CPVT both effort-induced polymorphic ventricular tachycardia and a mutant ryanodine receptor. It is also of note that a temperature-dependent dysfunction of the SCN5A gene for cardiac sodium channel has been characteristically observed in patients with Brugada syndrome (and Thr1620Met missense mutation). This increased temperature sensitivity could predispose some Brugada patients to life-threatening arrhythmias either during a febrile state or when body temperature increases during intense physical exertion.
- Intense static (isometric) exertion, such as lifting free weights, may prove to be adverse by inducing a Valsalva maneuver and dynamic left ventricular outflow obstruction in hypertrophic cardiomyopathy (HCM) (as well as the risk for traumatic injury in the event of impaired consciousness) or by increasing wall stress and weakening of the aortic media in Marfan syndrome, particularly if aortic dilatation is already present.
- Patients with diseases associated with impaired consciousness (e.g., syncope and near-syncope) are subject to considerably higher risk for traumatic injury while engaged in certain sports such as free weight and bench-pressing maneuvers, downhill skiing, diving, ice hockey, rock climbing, motorcycling, and horseback riding, and this factor should be taken into consideration in making recommendations to individual patients with GCVD.
- Although data are lacking, it is reasonable to specifically caution patients with GCVD, particularly those with catecholamine-sensitive or auditory-triggered arrhythmia syndromes such as long-QT syndrome (LQTS) and CPVT, against amusement park rides (e.g., roller coasters and thrill-related or frightening rides) because these are associated with intense stress and emotion due to sudden acceleration in heart rate and abrupt changes in centrifugal or centripetal forces.
- Paired athletic activities in which a second party may be at risk should the
  individual with GCVD suddenly incur bodily injury or impairment of
  consciousness and incapacitation (e.g., in recreational sports such as scuba
  diving and rock or mountain climbing). Water sports such as scuba diving or
  diving from platforms into pools are also generally unacceptable by virtue of
  their exposure of patients with GCVD (for whom syncope is a not-uncommon
  manifestation) to the risk of underwater drowning and reduced probability of
  rescue.
- Extreme sports (such as hang gliding and bungee jumping) are activities that are best avoided because they require the expenditure of particularly substantial physical energy and incur psychological demands that are

- exceedingly unpredictable, placing individuals with GCVD in compromised circumstances in which the likelihood of injury is substantial and the possibility of rescue from a traumatic or cardiovascular event is greatly reduced.
- Concomitant use of substances or compounds professed to promote enhanced physical performance but that also harbor the potential for adverse effects, particularly when associated with disease states or extreme environmental conditions (i.e., cocaine, anabolic steroids, or dietary and nutritional supplements such as ma huang, an herbal source of ephedrine [i.e., elemental ephedra] and a cardiac stimulant that is potentially arrhythmogenic).

# Table: Recommendations for the Acceptability of Recreational (Noncompetitive) Sports Activities and Exercise in Patients with GCVDs\*

Intensity level	HCM <sup>1</sup>	LQTS <sup>1</sup>	Marfan Syndrome <sup>2</sup>	ARVC	Brugada Syndrome
High					
Basketball					
Full court	0	0	2	1	2
Half court	0	0	2	1	2
Body building <sup>3</sup>	1	1	0	1	1
Ice hockey <sup>3</sup>	0	0	1	0	0
Racquetball/squash	0	2	2	0	2
Rock climbing <sup>3</sup>	1	1	1	1	1
Running (sprinting)	0	0	2	0	2
Skiing (downhill) <sup>3</sup>	2	2	2	1	1
Skiing (cross-country)	2	3	2	1	4
Soccer	0	0	2	0	2
Tennis (singles)					
Touch (flag) football	0	0	3	0	2
Windsurfing <sup>4</sup>	1	1	3	1	3
winasuring	1	0	1	1	1

Intensity level	HCM <sup>1</sup>	LQTS <sup>1</sup>	Marfan Syndrome <sup>2</sup>	ARVC	Brugada Syndrome
Moderate					
Baseball/softball	2	2	2	2	4
Biking	4	4	3	2	5
Modest hiking	4	5	5	2	4
Motorcycling <sup>3</sup>	3	1	2	2	2
Jogging	3	3	3	2	5
Sailing <sup>4</sup>	3	3	2	2	4
Surfing <sup>4</sup>	2	0	1	1	1
Swimming (lap) <sup>4</sup>	5	0	3	3	4
Tennis (doubles)	4	4	4	3	4
Treadmill/stationary bicycle	5	5	4	3	5
·	1	1	0	1	1
Weightlifting (free weights) <sup>3</sup> , <sup>5</sup>	3	3	3	2	4
Hiking					
Low					
Bowling	5	5	5	4	5
Golf	5	5	5	4	5
Horseback riding <sup>3</sup>	3	3	3	3	3
Scuba diving <sup>4</sup>	0	0	0	0	0
Skating <sup>6</sup>	5	5	5	4	5
Snorkeling <sup>4</sup>	5	0	5	4	4
Weights (non-free	4	4	0	4	4
weights)	5	5	5	5	5

Intensity level	HCM <sup>1</sup>	LQTS <sup>1</sup>	Marfan Syndrome <sup>2</sup>	ARVC	Brugada Syndrome
Brisk walking					

<sup>\*</sup>Recreational sports are categorized with regard to high, moderate, and low levels of exercise and graded on a relative scale (from 0 to 5) for eligibility with 0 to 1 indicating generally not advised or strongly discouraged; 4 to 5 indicating probably permitted; and 2 to 3 indicating intermediate and to be assessed clinically on an individual basis. The designations of high, moderate, and low levels of exercise are equivalent to an estimated >6, 4 to 6, and <4 metabolic equivalents, respectively.

# **Special Circumstances**

#### **Physical Education Class**

Issues related to recreational exercise often arise with regard to compulsory physical education classes in junior high or elementary school. In the former, the required levels of exercise in physical education classes vary considerably but often involve vigorous exertion and circumstances that are difficult to control. For example, although many components of such classes may be truly recreational, others clearly are not and can be regarded as competitive in nature (i.e., prolonged aerobic events such as the traditional timed 600-yard [or 400-meter] run and the President's Physical Fitness Award). On the other hand, in elementary school physical education class, sports activities may involve nothing more than innocent play.

<sup>&</sup>lt;sup>1</sup>Assumes absence of laboratory deoxyribonucleic acid (DNA) genotyping data; therefore, limited to clinical diagnosis.

<sup>&</sup>lt;sup>2</sup>Assumes no or only mild aortic dilatation.

<sup>&</sup>lt;sup>3</sup>These sports involve the potential for traumatic injury, which should be taken into consideration for individuals with a risk for impaired consciousness.

<sup>&</sup>lt;sup>4</sup>The possibility of impaired consciousness occurring during water-related activities should be taken into account with respect to the clinical profile of the individual patient. Barotrauma is a primary risk associated with the use of the scuba apparatus in Marfan syndrome.

<sup>&</sup>lt;sup>5</sup>Recommendations generally differ from those for weight-training machines (non-free weights), based largely on the potential risks of traumatic injury associated with episodes of impaired consciousness during bench-press maneuvers; otherwise, the physiological effects of all weight-training activities are regarded as similar with respect to the present recommendations.

<sup>&</sup>lt;sup>6</sup>Individual sporting activity not associated with the team sport of ice hockey.

Although in the view of some very conservative clinicians, it might seem most prudent for patients with GCVDs to largely avoid any involvement in school-structured physical education, the guideline developers recognize that profound personal and psychological stigmata can be associated with such selective, targeted, and often unnecessary gym class prohibition. Indeed, in some states, such children are placed in alternative health classes, which results in further ostracism by their peers. In the event that the patient and family judge it important, for social and peer-related reasons, to retain some level of participation and normalcy among peers (as is often the case), careful and detailed review of physical education class requirements should be undertaken by the parents in concert with school officials and their physician, during which the principles of safe recreational activities are agreed upon.

This process will define acceptable portions of the curriculum and exclude activities that can be regarded as intense or competitive (particularly those involving burst exertion), although it may be challenging to distinguish recreational from competitive activity under these circumstances. In this regard, it should be determined whether physical education instructors will agree to (or are capable of) monitoring the patient's activity level in accord with the medical recommendations. Therefore, the degree to which a child or adolescent with GCVD can or should participate in physical education class requires a large measure of individualization.

# **Implantable Cardioverter-Defibrillator**

A growing number of young patients with GCVDs are receiving implantable defibrillators for primary or secondary prevention of sudden death. Such patients may participate in a wide variety of noncompetitive and noncontact physical activities in concert with the exercise recommendations of the responsible electrophysiologist. However, the defibrillator itself may create some restrictions to these activities, particularly with regard to the possibility that bodily trauma may disrupt the lead system or that certain levels of physical exertion may trigger an inappropriate shock due to sinus tachycardia. Therefore, young patients with GCVD undergoing implantation of a cardioverter-defibrillator should receive a dual-chamber device with algorithms that improve discrimination between ventricular and supraventricular arrhythmias, thereby reducing the incidence of inappropriate interventions. Participation in intense exercise should not be advocated explicitly because of the presence of an implanted defibrillator and the antiarrhythmic protection afforded by the device.

## **CLINICAL ALGORITHM(S)**

None provided

# **EVIDENCE SUPPORTING THE RECOMMENDATIONS**

# TYPE OF EVIDENCE SUPPORTING THE RECOMMENDATIONS

The type of evidence supporting each recommendation is not specifically stated.

## BENEFITS/HARMS OF IMPLEMENTING THE GUIDELINE RECOMMENDATIONS

## **POTENTIAL BENEFITS**

- The health benefits of exercise at all ages have been emphasized repeatedly and promoted as a national public health agenda. Certainly, there is substantial evidence that considerable medical advantage is derived from even regular moderate exercise and fitness, such as improvement in aerobic power and maximum oxygen uptake, blood lipid levels and glucose tolerance, as well as enhanced self-assurance, a sense of psychological and physical well-being, and improved overall quality of life. Undoubtedly, similar benefits from regular exercise probably also accrue in a young patient population with genetic cardiovascular diseases (GCVDs).
- In addition, recent recognition in the United States that obesity is an emerging major health problem in young people has focused attention on the importance of regular exercise as a weight loss and maintenance strategy in adults. In general terms, the guideline panel believes these principles are also relevant to young patients with GCVD.

## **POTENTIAL HARMS**

The panel found it useful to express specific exercise recommendations in terms of those activities that should be avoided by patients with clinically diagnosed genetic cardiovascular disease (GCVD). See the "Major Recommendations" field for a discussion of these activities.

## **QUALIFYING STATEMENTS**

## **QUALIFYING STATEMENTS**

- These recommendations are not intended to be (and cannot be) rigid dicta but rather should be viewed as general guidelines, and not specific standards, that allow sufficient latitude to the managing physician for individual clinical judgment.
- The present recommendations assume the absence of important limiting cardiac symptoms, as well as an unequivocal cardiac diagnosis previously made on the basis of clinically overt features. Therefore, young people with negligible manifestations of disease, who may harbor only preclinical (i.e., molecular) evidence of a particular disease-causing mutation predisposing to genetic cardiovascular disease (GCVD), are excluded. Furthermore, at present, it is unresolved as to whether genotype-positive/phenotype-negative individuals warrant any restrictions from either recreational or competitive sports.
- Issues directly related to preparticipation screening for cardiovascular disease are beyond the scope of this document. The panel also recognizes that the present recommendations, in some instances, may be reasonably applied to patients older than 40 years (in the absence of important comorbidity), as well as to employment- and occupation-related activities involving physically vigorous and intense lifestyles (e.g., firemen and other emergency personnel).

- The panel operated under several major conceptual premises in formulating the recommendations. First, vigorous physical activity may trigger sudden death in susceptible individuals with underlying heart disease. Undoubtedly, there are other potential mechanisms, because sudden death is also known to occur with modest or sedentary activity or even during sleep, or to be triggered by abrupt or loud noises (such as in long-OT syndrome [LOTS]), but these variables remain largely undefined. Second, the vast majority of sudden deaths associated with GCVDs are due to primary ventricular tachyarrhythmias, although in Marfan syndrome sudden death is usually secondary to aortic dissection and rupture; however, arrhythmic sudden death has also been reported. Third, the risk for cardiac events associated with exercise in patients with GCVDs is, in theory, amenable to interventional strategies and therefore avoidable to a large extent. This latter tenet is fundamental to the present document because it supports the principle that sudden death may be prevented (or the risk substantially reduced) through lifestyle management and restriction of exercise or through the use of implantable devices.
- Implementation of exercise recommendations ultimately depends in large measure on the interaction between physician and patient. It may often be necessary for clinicians to individualize exercise recommendations for particular patients, the specific GCVD involved, and the physical activity under consideration. These clinical decisions regarding the structure of exercise programs are also unavoidably influenced by liability issues and concerns, the possibility that physician recommendations may be ignored by some patients, and the variable tolerance for sudden death risk among patients and their families.
- The panel found it difficult to design recommendations that rely on obtaining truly quantitative measurements dependent on monitoring, such as maximum heart rate or metabolic equivalents with exercise, given the diverse sporting disciplines involved, the multitude of variables that impact these activities, and the impracticality of making accurate assessments during athletic activity.
- In establishing a recreational exercise program for patients with known GCVDs, physician deviation from consensus guidelines in a particular individual case does not necessarily constitute medical malpractice if consistent with acceptable medical practices under the circumstances. The controlling legal issue is whether adherence to (or deviation from) consensus guidelines is consistent with reasonable, customary, and accepted medical practice in an individual patient. Consistent with this legal principle, the medical recommendations set forth in this document should not and cannot be viewed as inflexible doctrine but rather should allow sufficient latitude to the managing physician without precluding individual clinical judgment. Compliance with these recommendations is evidence that a physician has satisfied these legal requirements and may form the basis of a successful defense to allegations of malpractice.

## **IMPLEMENTATION OF THE GUIDELINE**

## **DESCRIPTION OF IMPLEMENTATION STRATEGY**

An implementation strategy was not provided.

# INSTITUTE OF MEDICINE (IOM) NATIONAL HEALTHCARE QUALITY REPORT CATEGORIES

## **IOM CARE NEED**

Living with Illness

## **IOM DOMAIN**

Effectiveness Patient-centeredness

## **IDENTIFYING INFORMATION AND AVAILABILITY**

## **BIBLIOGRAPHIC SOURCE(S)**

Maron BJ, Chaitman BR, Ackerman MJ, Bayes de Luna A, Corrado D, Crosson JE, Deal BJ, Driscoll DJ, Estes NA 3rd, Araujo CG, Liang DH, Mitten MJ, Myerburg RJ, Pelliccia A, Thompson PD, Towbin JA, Van Camp SP. Recommendations for physical activity and recreational sports participation for young patients with genetic cardiovascular diseases. Circulation 2004 Jun 8;109(22):2807-16. [86 references] PubMed

#### **ADAPTATION**

Not applicable: The guideline was not adapted from another source.

## **DATE RELEASED**

2004 Jun 8

# **GUIDELINE DEVELOPER(S)**

American Heart Association - Professional Association

## **SOURCE(S) OF FUNDING**

American Heart Association

#### **GUIDELINE COMMITTEE**

Working Groups of the American Heart Association Committee on Exercise, Cardiac Rehabilitation, and Prevention

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# FINANCIAL DISCLOSURES/CONFLICTS OF INTEREST

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## **GUIDELINE STATUS**

This is the current release of the guideline.

#### **GUIDELINE AVAILABILITY**

Electronic copies: Available from the American Heart Association Web site:

- HTML Format
- Portable Document Format (PDF)

Print copies: Available from the American Heart Association, Public Information, 7272 Greenville Ave, Dallas, TX 75231-4596; Phone: 800-242-8721

# **AVAILABILITY OF COMPANION DOCUMENTS**

None available

#### **PATIENT RESOURCES**

None available

## **NGC STATUS**

This NGC summary was completed by ECRI on October 12, 2004. The information was verified by the guideline developer on October 19, 2004.

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